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# Multiple Inflammatory Fibrosarcoma of the Abdominal Cavity in a Child

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Inflammatory fibrosarcoma is a rare condition in childhood. In the abdominal location, its behaviour is often aggressive and potentially metastasizing. We report a case of a 3-year-old female with abdominal inflammatory fibrosarcoma who relapsed after 1 month from radical surgery. Chemotherapy was ineffective, and we registered a brief stabilisation of disease only with α-IFN. Our case confirms the potential malignancy of this tumour and its resistance to treatment. It is noteworthy that the therapy with α-IFN improved the quality of life in this child for 4 months. 

**Key words:** Fibrosarcoma, chemotherapy, α-IFN

## INTRODUCTION

Inflammatory pseudotumor is a rare condition in childhood reported as plasma cell granuloma, fibrohistiocytoma, plasma-cell histiocytoma, or inflammatory myofibrohistiocytic proliferation [1–2]. Usually it simulates a true neoplasm arising in the lung and less frequently in the mediastinum, liver, retroperitoneum, mesentery, urinary bladder, meninges, cerebrum, and nodes [3].

In 1991, Meis and Enzinger [3] reported experience in the treatment of inflammatory fibrosarcoma (IF) of the abdominal cavity and suggested that this tumor is locally aggressive and potentially metastasizing and should be labelled as sarcoma. We report a case of abdominal inflammatory fibrosarcoma with a highly aggressive local behaviour. Chemotherapy was ineffective, and a brief arrest of progression was obtained only with α IFN.

## CASE REPORT

A 3-year-old female was admitted to Gaslini Children's Hospital in April 1994 with a 1-month history of fever, anemia, and abdominal mass. On physical examination she appeared chronically ill, pale, and in poor general condition. A firm mass was palpated in the right upper quadrant and another mass in the pelvic region.

A CT scan revealed a large solid mass measuring ~8 × 5 cm displacing the anterior wall of the stomach and in contact with liver and another smaller mass upon the bladder (Fig. 1). The mass presented an inhomogenous structure with a central necrotic area without calcifications. Laboratory data included leukocyte count 16.0 × 10/mm$^3$ with 45% lymphocytes; the hemoglobin was 8,2 g/dl, and the platelet count was 480 × 10$^9$/l. On peripheral smear, the RBC appeared microcytic and hypochromic. Other studies included serum iron 10 ng/dl, IgG 1080 mg/dl, IgA 400 mg/dl, IgM 200 mg/dl; LDH 295 UI/l; the urinary catecholamine excretion and α-feto protein were normal for age. The bone marrow aspirate was normal.

At surgery an 8 × 5 cm solid tumour, well circumscribed and attached to the omentum was found. It was easily and completely excised. The cut surfaces were whitish-grey to pale tan in color with different areas of necrosis.

Microscopically, two different cellular constituents were identified in the mass: spindle cells and mononuclear inflammatory cells [3]. Numerous mature plasma cells and small lymphocytes were among the spindle cells. Lymphoid aggregates with germinal centers were found locally. The histological features were typical of IF. Two weeks after primary surgery, the epigastric mass was also removed (Fig. 2). At laparotomy, a large mass between the stomach and liver was disclosed. A complete excision of the mass was performed, including a partial resection of both liver (left lobe) and stomach. The exploration of the abdominal cavity was negative. Both lesions showed the same histological features.

In all sections the morphological picture was composed

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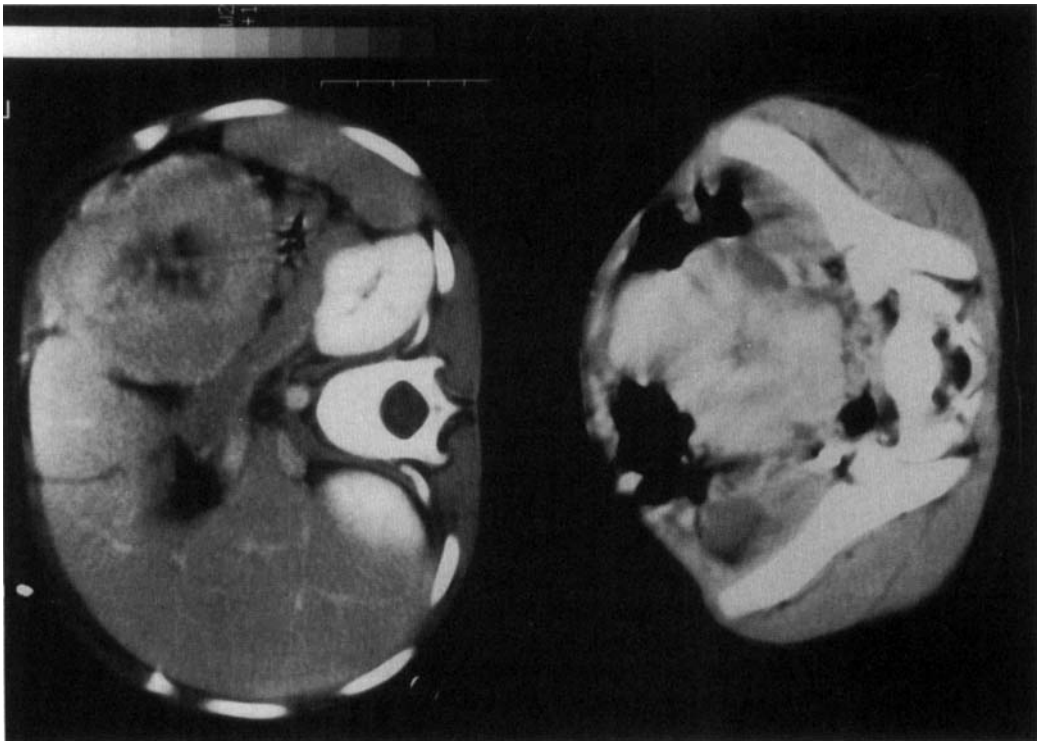

**Fig. 1.** Left: Upper abdominal CT, that shows a solid intraperitoneal mass, with a low density central part, anterior to the tail of the pancreas. Right: solid irregular mass in the pelvis of the same patient.

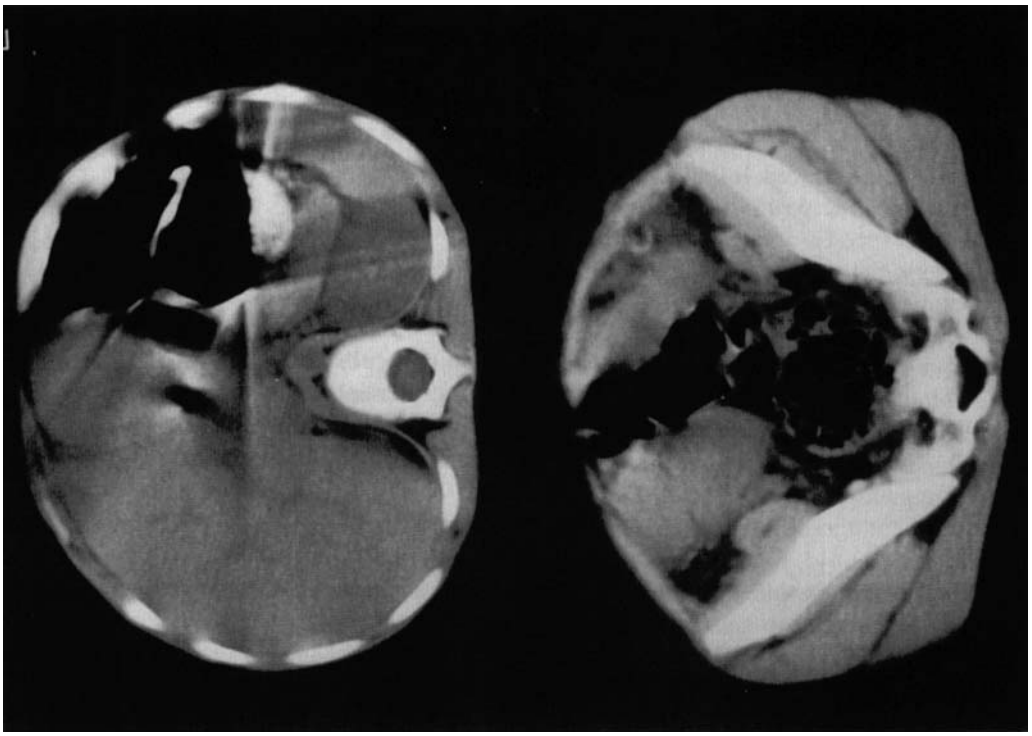

**Fig. 2.** Postoperative CT: normal findings in the upper abdomen. Minimal residual pathologic tissue in the pelvis.

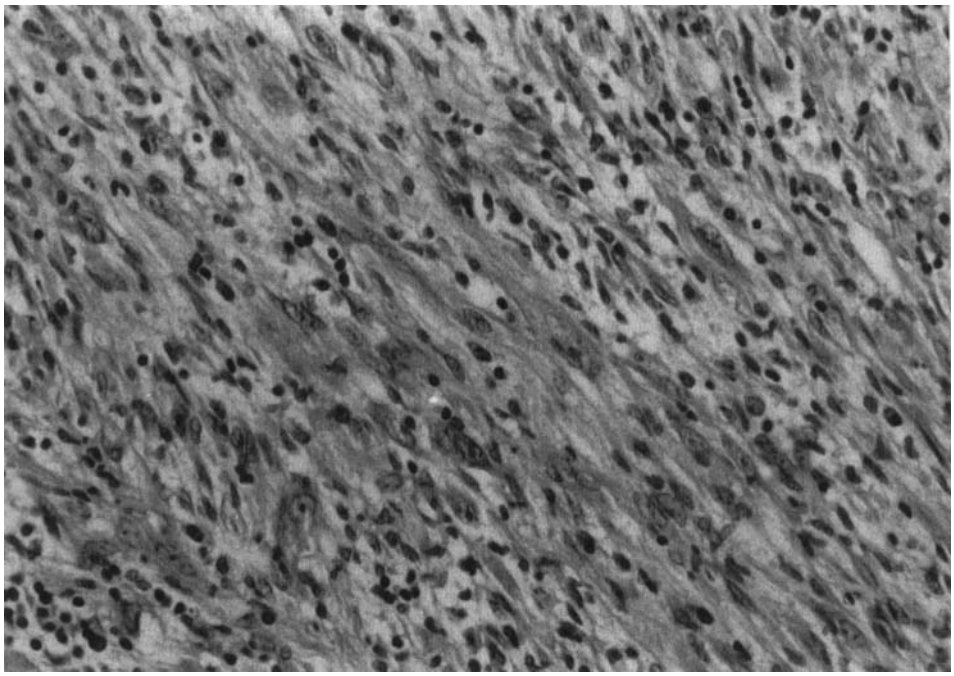

**Fig. 3.** Atypical spindle cell proliferation intermingled with lymphocytes and plasma cells.

of atypical spindle cells associated with an intense inflammatory infiltrate (Fig. 3) [3–5]. Spindle cells were arranged in small interlacing fascicles or whorls. These cells showed oval or irregular nuclei with nucleoli and a large amount of basophilic cytoplasm. Larger cells with huge inclusion-like nucleoli were observed. Mitotic figures were not frequent. The inflammatory infiltrate was prominent and composed mostly of plasma cells and lymphocytes with rare leukocytes. A few lymphoid follicles with germinal centers were observed. Immunohistochemical study showed cytoplasmic positivity in spindle cells to vimentin and muscle-specific actin.

After second surgery, the patient's general conditions improved and fever disappeared. A CT scan performed 1 month later showed a pelvic relapse (Fig. 4). The patient underwent a third-look surgery, disclosing multiple nodular lesions in the colon, mesentery, and rectum. Biopsy of two of these lesions was performed, and histology confirmed the diagnosis of IF.

Considering the aggressiveness of this disease, we started unsuccessful chemotherapy with cyclophosphamide—VP 16 and corticosteroid, followed after 3 weeks by epirubicin and carboplatin. Despite this treatment, disease progressed, and we decided to start administration of α-IFN 3.000.000 U/m$^2$ 3 days/week to exceed the capacity of α-IFN to stop inflammatory cells and angiogenesis. The child improved, fever disappeared, and CT and US scans showed a stable disease for 4 months; then the disease progressed. At present the patient is in a terminal status at home with analgesic therapy.

## CONCLUSION

The inflammatory fibrosarcoma and inflammatory pseudotumor must be considered two distinct clinical pathological entities [3,4]. The former is very rare in children and behaves aggressively, especially in the abdomen where contiguous organs are frequently involved. Surgical excision is the treatment of choice, but local recurrence is frequent. The occurrence of distant metastases in lungs or brain reported in 10% of cases further confirms the potential malignancy of this neoplasm.

Most IF are located in the mesentery, retroperitoneum, and omentum, and multiple nodules are observed at presentation in almost 30% of reported cases.

Since the histology is characterized by a proliferation of spindle cells associated with an inflammatory infiltrate, the differential diagnosis includes retroperitoneal fibrosis, inflammatory malignant fibrous histiocytoma, xanthogranuloma and inflammatory pseudotumour. The distinction between IF and inflammatory pseudotumor can be difficult, the former showing atypical cytological features in the spindle cell component [2–3].

As suggested by Meis and Enzinger [3], the tumor arising in the abdomen presents an aggressive behaviour with transmural and regional invasion, multiple peritoneal implants, local recurrence, and distant metastases. Few cases reported in literature were treated with radiotherapy or chemotherapy without a clear benefit [3,5].

Our case confirms the aggressive behaviour of this tumour and its resistance to treatment. However, it is

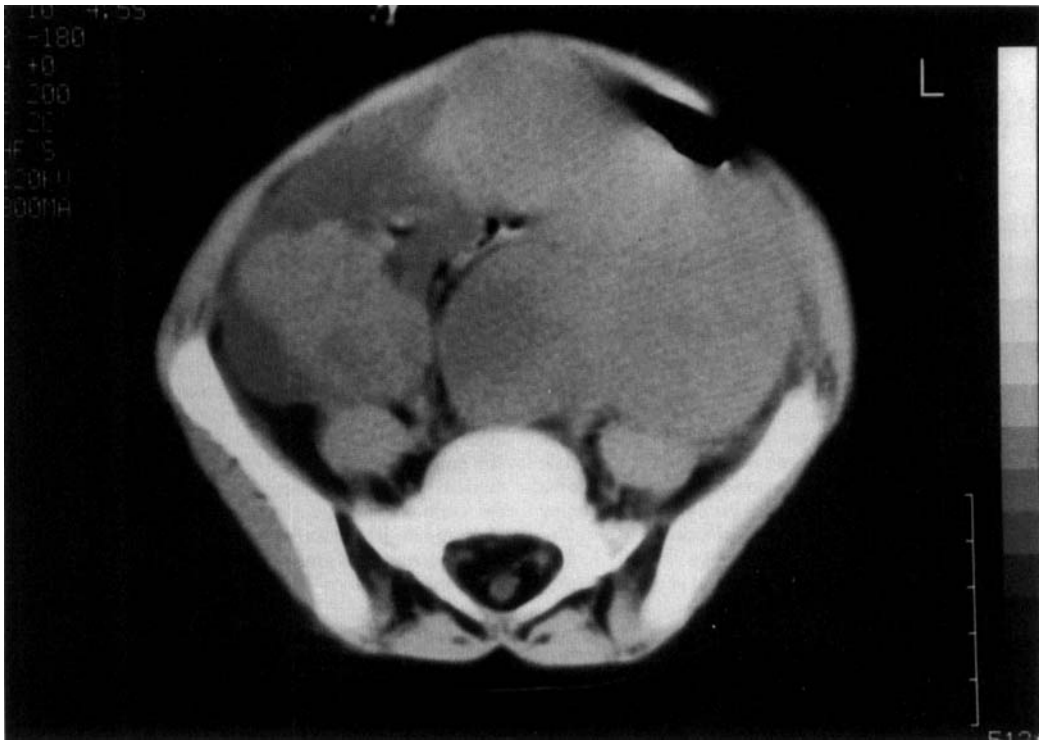


**Fig. 4.** CT performed after 1 month from surgery shows a large relapse in the pelvis.

interesting to note that during therapy with α-IFN we registered stable disease for 4 months with improvement of the quality of life.

## ACKNOWLEDGMENTS

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